Clues for discovering Cause of Status Epilepticus

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Outline

1. Definition of Status Epilepticus
2. Classification of Status Epilepticus
3. Common cause of Status Epilepticus
4. Less common cause of Status Epilepticus
5. Conclusion

Status Epilepticus

Definition of Status Epilepticus

“A seizure that lasts longer than 5 minutes, or having more than 1 seizure within a 5 minutes period, without returning to a normal level of consciousness between episodes”

The Neurocritical Care Society guidelines from 2012

Table 1. Proposed classification of seizure types according to their semiology, along two taxonomic criteria: motor symptoms and impairment of consciousness

- With prominent motor symptoms
  - Convulsive SE (syn.: tonic–clonic SE)
  - Myoclonic SE (*prominent epileptic myoclonic jerks*)
  - Focal motor (*including EPC*)
  - Tonic SE
  - Hyperkinetic SE
- Without prominent motor symptoms (i.e., NCSE)
  - NCSE with coma
  - NCSE without coma
    - Generalized
    - Focal
- Boundary syndromes
  - Epileptic encephalopathy
  - Acute forms of coma with status-like EEG pattern
  - Epileptic behavioral disturbance and psychosis
  - Confusional states, or delirium with epileptiform EEG changes

Table 2. Categories of NCSE, classified according to the degree of disturbed consciousness

- NCSE with coma
- NCSE without coma
  - Generalized
    - Typical absence status
    - Atypical absence status
    - Myoclonic absence status
  - Focal
    - Aura continua
      - With vegetative symptoms
      - With sensory symptoms
      - With visual symptoms
      - With olfactory symptoms
      - With gustatory symptoms
      - With emotional symptoms
    - Aphasic SE
    - SE with dyscognitive symptoms

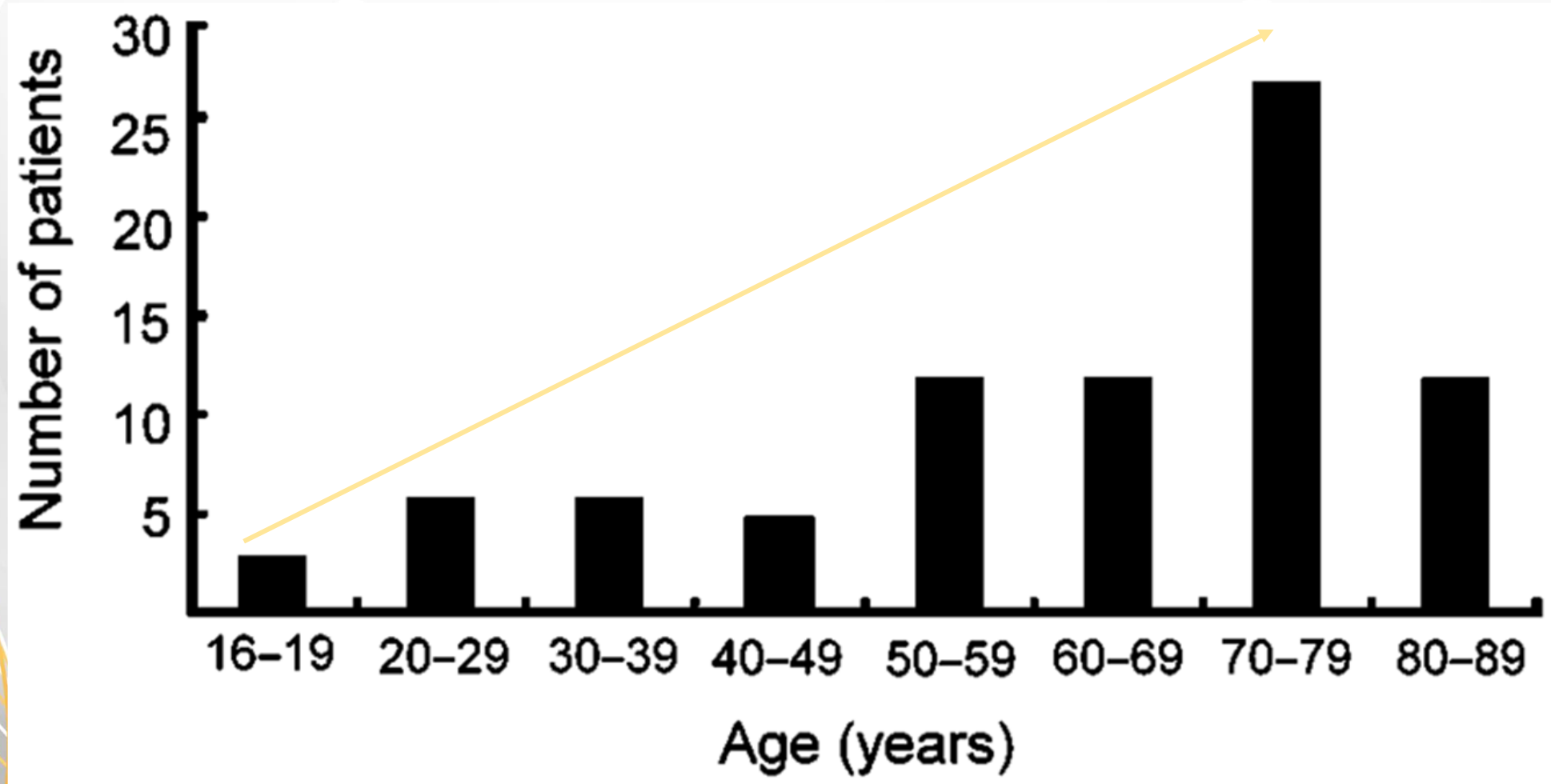
Eugen Trinka, Epilepsia, 53(Suppl. 4):127–138, 2012

Table 1. Clinical Features of Nonconvulsive Status Epilepticus[7]

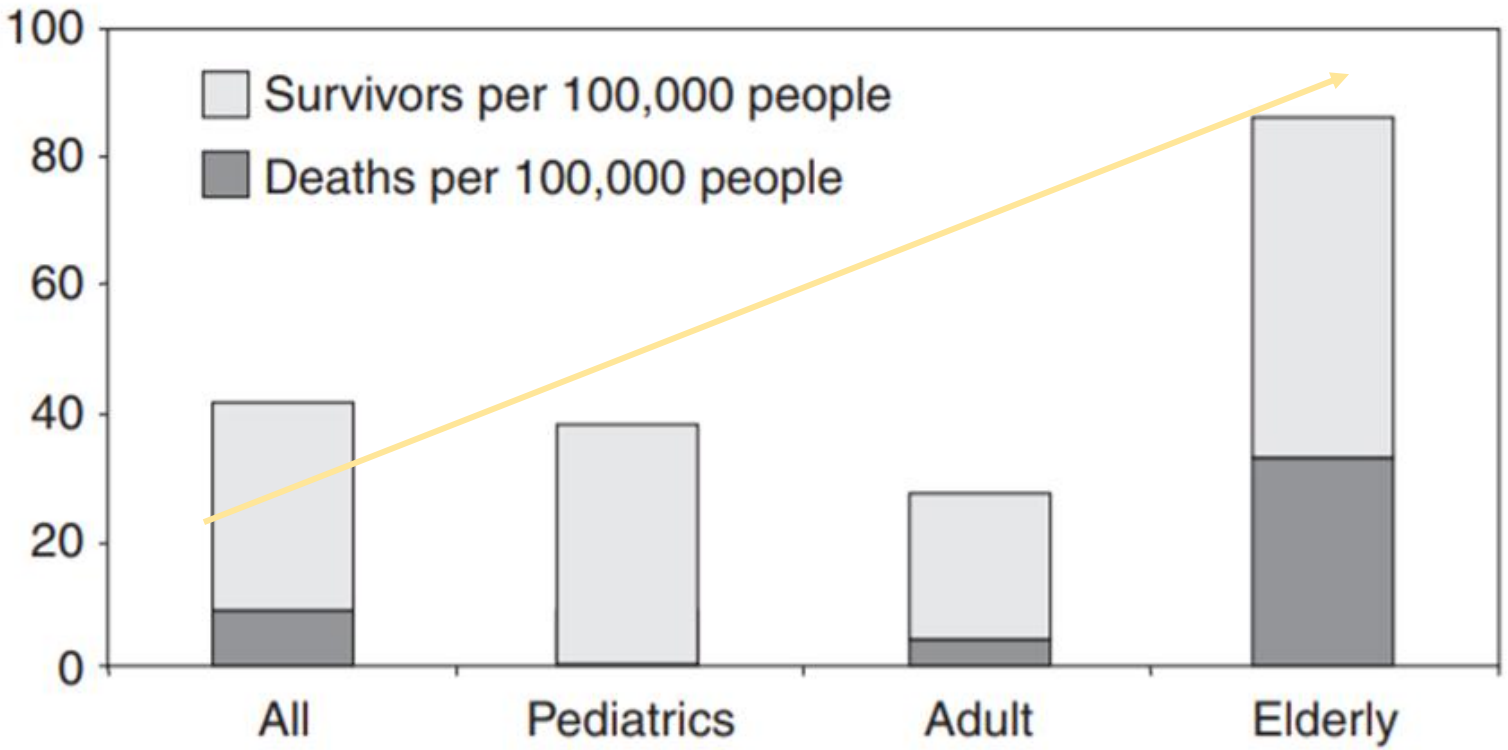
- Altered mental status (82%)
  - Confusion (49%)
  - Coma (22%)
  - Lethargy (21%)
  - Memory loss (8%)
- Speech disturbance (15%)
- Myoclonus (13%)
- Unusual behavior (11%)
- Anxiety, agitation, and delirium (8%)
- Extrapyramidal signs (7%)
- Hallucinations (6%)

Sarice Bassin, Critical Care 2002

The age distributions of 83 de novo SE patients.



M.-H. TSAI. Q J Med 2009



A graphical representation of mortality and incidence for four population groups. These data are adapted from *Neurology* [19] and *Epilepsia* [21].

Sarice Bassin, Critical Care 2002

Major factors increased risk of mortality and morbidity in SE

Age > 60 years

Long Duration of SE

Etiologies

D

A

E

Towne et al.,1994; DeLorenzo et al., 1996; Wu et al., 2002; Rossetti et al., 2006

Table 1

Data on the etiology of status epilepticus in an urban hospital-based practice

Etiology	Percent of cases
AED non-compliance	26
Alcohol related	24
Drug toxicity	10
CNS infection	8
Cerebral tumor	6
Trauma	5
Refractory epilepsy	5
Stroke	4
Metabolic abnormalities	4
Cardiac arrest	4
Idiopathic	5

AED, anti-epileptic drugs; CNS, central nervous system. Data adapted from *Neurology* [16].

Table 2

Data on the etiology of status epilepticus in a hospital and community

Etiology	Percent of Cases
Withdrawal of anticonvulsants	25
Cerebrovascular disease	23
Remote symptomatic	19
Alcohol withdrawal	15
Metabolic disorders	13
Hypoxia	12
Infectious disorders	8
Tumors	5
Anoxia	4
Trauma	3
Hemorrhage	2
Drug overdose	2
Idiopathic	4

Data adapted from Churchill Livingstone [17].

Sarice Bassin, Critical Care 2002

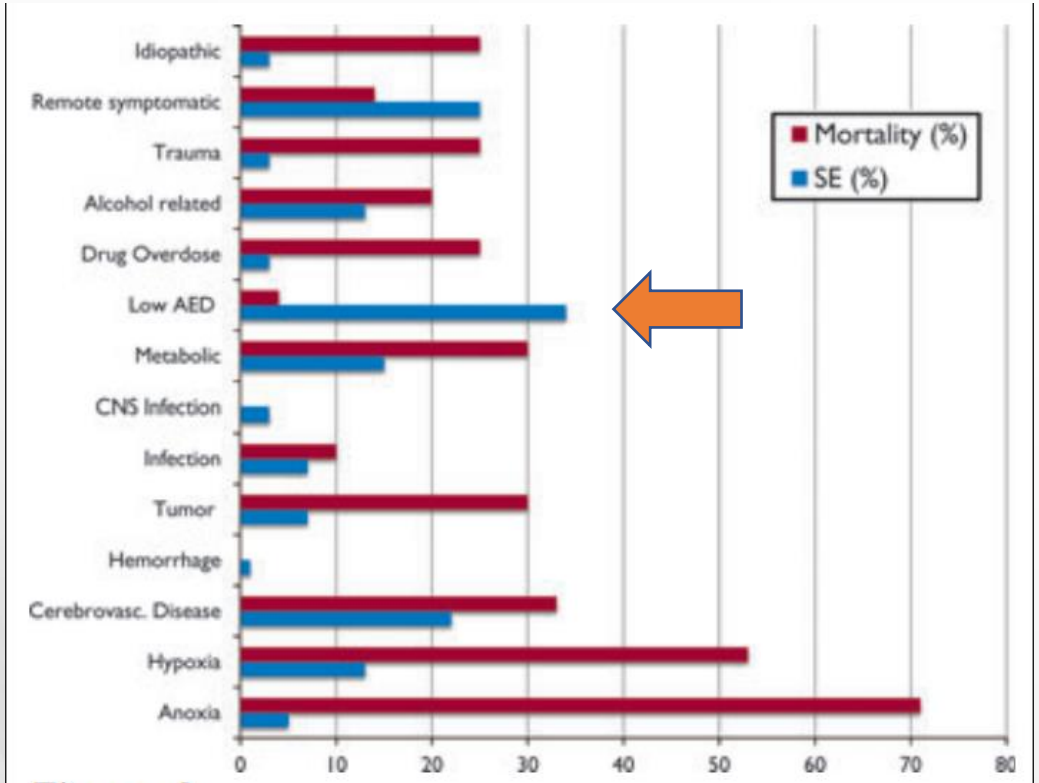


Figure 1.

Etiology of status epilepticus in adults, with associated mortality for each category. Based on data from DeLorenzo et al., 1995. AED, antiepileptic drugs; CNS, central nervous system.

Epilepsia © ILAE

Causes of Status Epilepticus in Adults

METABOLIC DISTURBANCES
Hepatic encephalopathy
Hypocalcemia
Hypoglycemia or hyperglycemia
Hyponatremia
Uremia

INFECTIOUS PROCESSES
Central nervous system abscess
Encephalitis
Meningitis

WITHDRAWAL SYNDROMES
Alcohol
Antiepileptic drugs
Baclofen
Barbiturates
Benzodiazepines

CENTRAL NERVOUS SYSTEM LESIONS
Acute hydrocephalus
Anoxic or hypoxic insult
Arteriovenous malformations
Brain metastases
Cerebrovascular accident
Eclampsia
Head trauma: acute and remote
Intracerebral hemorrhage
Neoplasm
Posterior reversible leukoencephalopathy

INTOXICATION
Bupropion
Camphor
Clozapine
Cyclosporine
Flumazenil
Fluoroquinolones
Imipenem
Isoniazid
Lead
Lidocaine
Lithium
MDMA
Metronidazole
Synthetic cannabinoids
Theophylline
Tricyclic antidepressants

Source : Rosens Emergency Medicine Concepts and Clinical Practice, 2-Volume Set, 9th Edition

Table 6. Causes of epilepsia partialis continua (according to Tan et al., 2010)

Age period	EPC type 1 (static cause)	Diagnostic test	EPC type 2 (progressive cause)	Diagnostic test
	(Ketotic) hyperglycemia			
Adults	Cerebrovascular disorders (stroke; intracranial bleeding, cerebral venous thrombosis, vasculitis)		Adult-onset Rasmussen's syndrome	Cerebrospinal fluid oligoclonal banding, immunoglobulin G index
	Nonketotic (ketotic) hyperglycemia	Serum glucose	Creutzfeldt-Jakob disease	14-3-3 protein in cerebrospinal fluid
	Focal cortical dysplasia		Myoclonus epilepsy with ragged red fibers (MERRFs)	Serum and cerebrospinal fluid lactate, muscle biopsy, mitochondrial DNA
	Paraneoplastic limbic encephalopathy	Cerebrospinal fluid study, chest computed tomography, anti-Hu test	Kuf's disease	Skin or rectal mucosal biopsy
	Neoplasms			
	Tuberculous meningitis (tuberculoma)	Cerebrospinal fluid study, chest XR, tuberculin skin test		
	(Tick-borne) encephalitis	Cerebrospinal fluid study, serologic test for virus		
	Autoimmune thyroid encephalopathy	Thyroid function tests, antithyroglobulin antibody, antimicrosomal antibody		
	Behcet disease	Neuroimaging, recurrent oral and genital ulceration, skin lesions, HLA-B5 positivity		
	Sjögren syndrome	Hypergammaglobulinemia positive antinuclear antibody, anti-SSA, SSB, rheumatoid factor		
	Multiple sclerosis	Cerebrospinal fluid oligoclonal banding Immunoglobulin G index		
	HIV encephalopathy	Serologic test for HIV		

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Uncommon Causes of Status Epilepticus

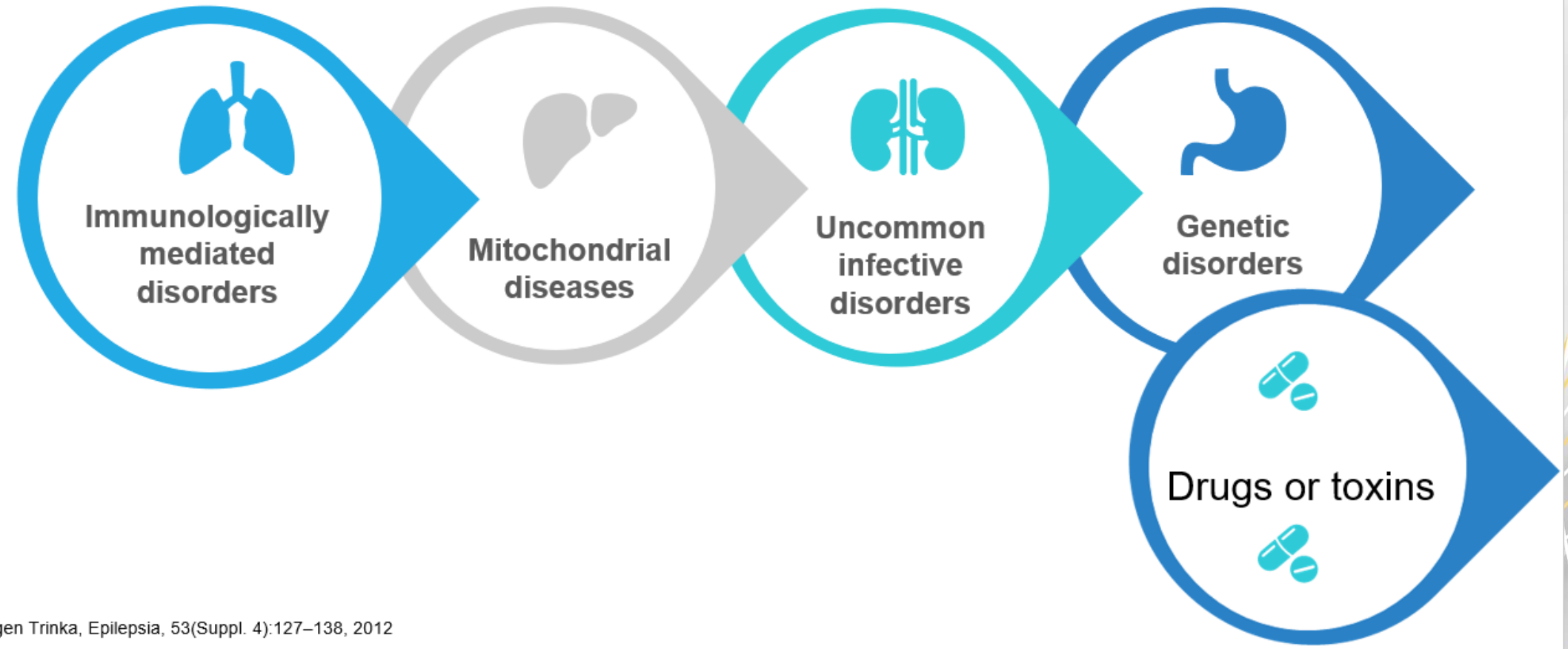

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Table 4. Immunologic disorders causing status epilepticus

Paraneoplastic encephalitis
Hashimoto encephalopathy
Anti-NMDA-receptor encephalitis
Anti-VGKC-receptor encephalitis
Rasmussen encephalitis
Cerebral lupus
Adult-onset Still disease
Anti-GAD antibody associated encephalitis
Goodpasture syndrome
Thrombotic thrombocytopenic purpura
Antibody-negative limbic encephalitis

NMDA, *N*-methyl-D-aspartate; GAD, glutamate acid decarboxylase.

Table 7. Mitochondrial diseases causing status epilepticus

Alpers disease
Occipital lobe epilepsy/mitochondrial spinocerebellar ataxia and epilepsy (MSCAE)
Mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes (MELAS)
Leigh syndrome
Myoclonic encephalopathy with ragged red fibers (MERRF)
Neuropathy, ataxia, and retinitis pigmentosa (NARP)

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Table 8. Uncommon infectious disease causing status epilepticus

Atypical bacterial infections	Viral infections	Prion disease	Other infections
Bartonella/cat-scratch disease	HIV and HIV-related infections	Creutzfeldt-Jakob disease	Paracoccidioidomycosis
Coxiella burnett (Q fever)	West Nile encephalitis		Paragonimiasis
Neurosyphilis	JC virus (progressive multifocal leukoencephalopathy)		Mucormycosis
Scrub typhus	Parvovirus B19		
Shigellosis	Varicella encephalitis		
Mycoplasma pneumonia	Subacute sclerosing panencephalitis		
Chlamydophilapsittaci	Measles encephalitis		
	Rubella encephalitis		
	Rous sarcoma virus		
	(RSV) associated SE		
	Polioencephalomyelitis		
	St. Louis encephalitis		

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Table 9. Status epilepticus due to genetic diseases

Chromosomal aberrations	Inborn errors of metabolism	Malformations of cortical development	Neurocutaneous syndromes	Others
Ring chromosome 20	Porphyria	Focal cortical dysplasias	Sturge-Weber syndrome	Rett's syndrome
Angelman syndrome	Menke's disease	Hemimegalencephaly	Tuberous sclerosis	Dravet syndrome and *SCN1A* gene mutation spectrum
Wolf-Hirschhorn syndrome	Wilson's disease	Polymicrogyria		Migrating partial seizures in infancy
Fragile X syndrome	Alexander's disease	Heterotopias		Pyeridoxine dependency
X-linked mental retardation syndrome	Gobalamin C/D deficiency	Schizencephaly		Familial hemiplegic migraine
Ring chromosome 17	Ornithine transcarbamylase (OTC) deficiency			Lafora's disease
	Hyperprolinemia			Dentato-rubro-pallido-luysian atrophy
	Maple-syrup urine disease			Infantile-onset spinocerebellar ataxia
	3-Methylcrotonyl CoA carboxylase deficiency			Wrinkly-skin syndrome
	Lysinuric protein intolerance			Neurocutaneous melanomatosis
	Hydroxyglutaric aciduria			Neuroserpin mutation
	Metachromatic leukodystrophy			Wolfram syndrome
	Kuf's disease			Autosomal recessive hyperekplexia
	Late infantile ceroid lipofuscinosis			Cockayne syndrome
	Beta-ureidopropionase deficiency			Cerebral autosomal dominant arterio-pathy with subcortical infarcts and leuko encephalopathy (CADASIL)
	3-Hydroxyaxyl CoA dehydrogenase deficiency			Jeavons syndrome
	Carnitine palmitoyltransferase			Robinow syndrome
	Succinic semildehyde dehydrogenase deficiency			*LYK5* mutation
				MECP2 mutation
				Malignant hyperpyrexia

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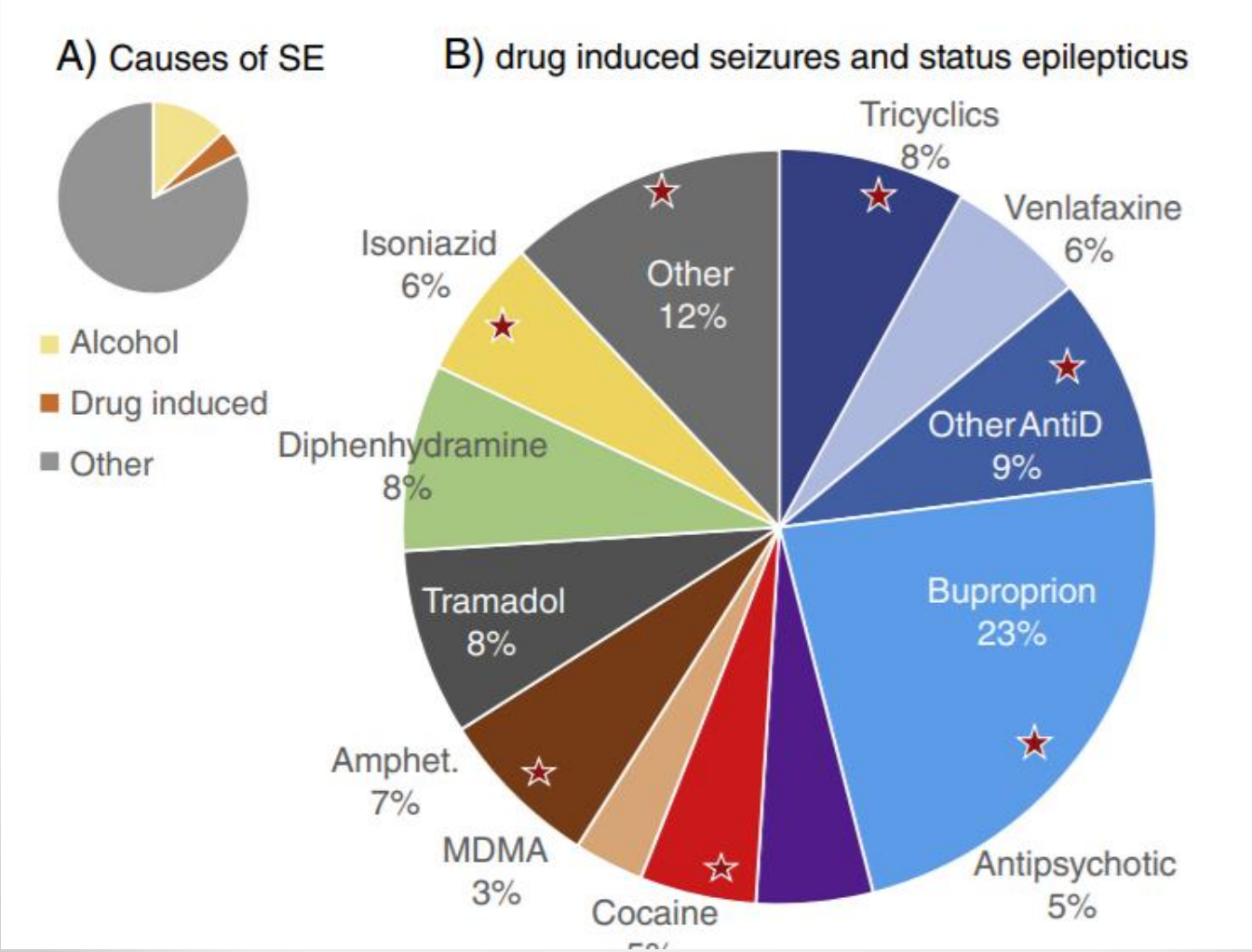

Hannah R. Cock. Epilepsy & Behavior 2015

Common cause of SE in NIT

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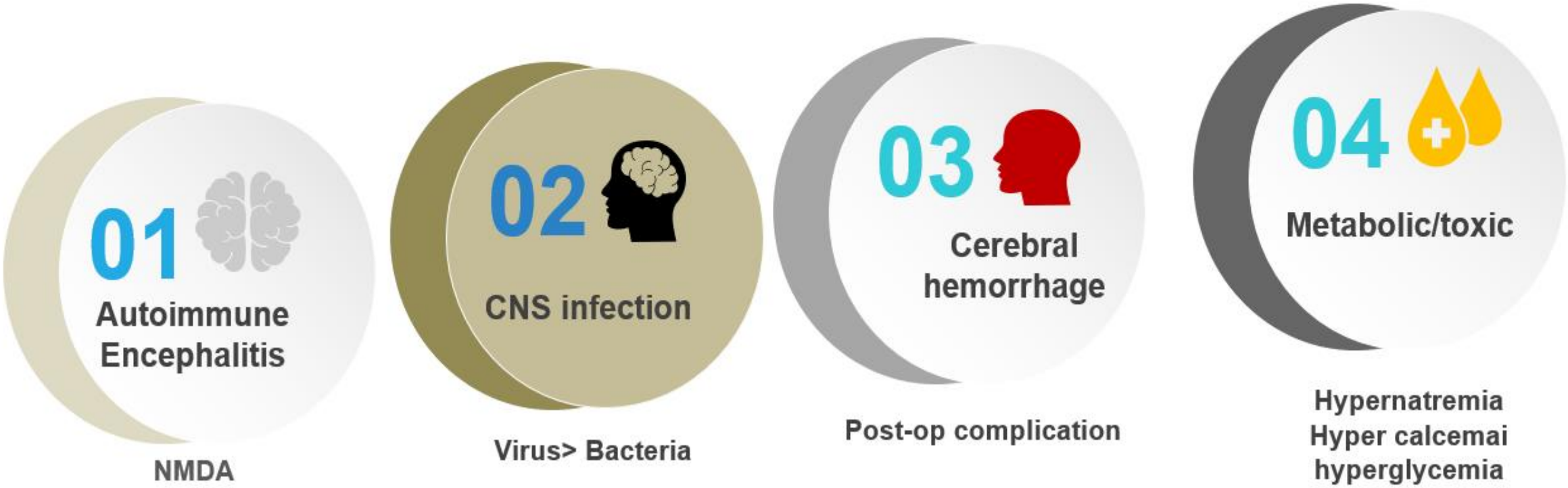
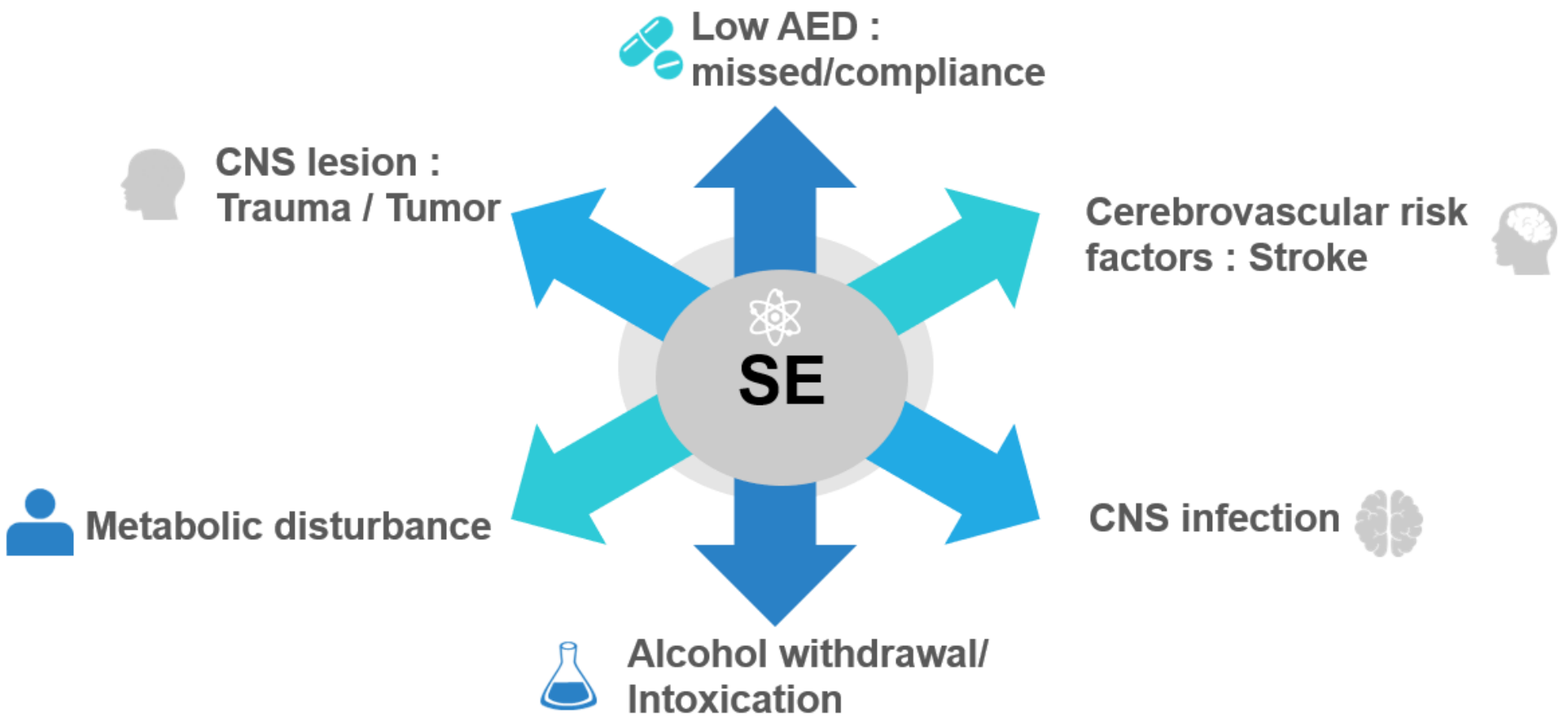
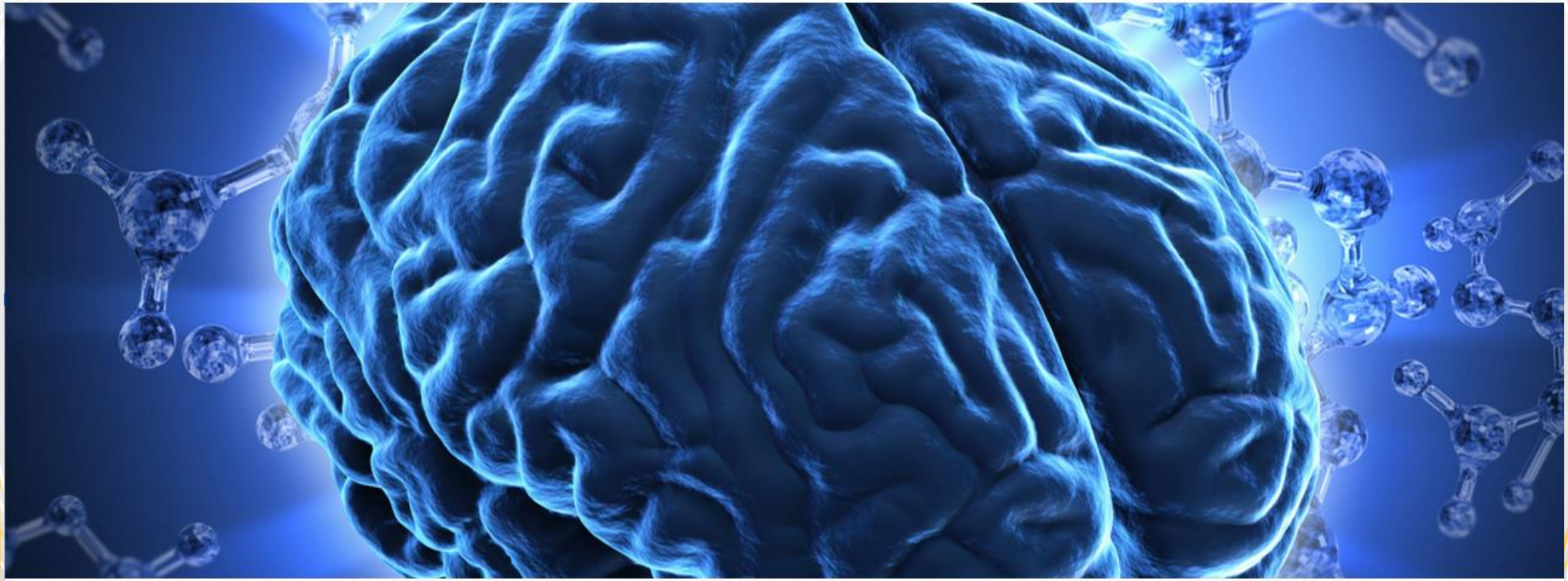


Conclusion


Low AED :
missed/compliance
CNS lesion :
Trauma / Tumor
Cerebrovascular risk
factors : Stroke
SE
Metabolic disturbance
CNS infection
Alcohol withdrawal/
Intoxication



Thank you